

Aortic valve disease

Aortic stenosis

Aetiology and pathophysiology

The likely aetiology depends on the age of the patient ([Box 18.105](#)). In congenital aortic stenosis, obstruction is present from birth or becomes apparent in infancy. With bicuspid aortic valves, obstruction may take years to develop as the valve becomes fibrotic and calcified. The aortic valve is the second most frequently affected by rheumatic fever and, commonly, both the aortic and mitral valves are involved. In older people, a structurally normal tricuspid aortic valve may be affected by fibrosis and calcification, in a process that is histologically similar to that of atherosclerosis affecting the arterial wall. Haemodynamically significant stenosis develops slowly, typically occurring at 30–60 years in those with rheumatic disease, 50–60 in those with bicuspid aortic valves and 70–90 in those with degenerative calcific disease.



18.105 Causes of aortic stenosis

Infants, children, adolescents

- Congenital aortic stenosis
- Congenital subvalvular aortic stenosis
- Congenital supra-annular aortic stenosis

Young adults to middle-aged

- Calcification and fibrosis of congenitally bicuspid aortic valve
- Rheumatic aortic stenosis

Middle-aged to elderly

- Senile degenerative aortic stenosis
- Calcification of bicuspid valve
- Rheumatic aortic stenosis

Cardiac output is initially maintained at the cost of a steadily increasing pressure gradient across the aortic valve. The LV becomes increasingly hypertrophied and coronary blood flow may then be inadequate; patients may therefore develop angina, even in the absence of concomitant coronary disease. The fixed outflow obstruction limits the increase in cardiac output required on exercise. Eventually, the LV can no longer overcome the outflow tract obstruction and pulmonary oedema supervenes. In contrast to patients with mitral stenosis, which tends to progress very slowly, those with aortic stenosis typically remain asymptomatic for many years but deteriorate rapidly when symptoms develop, and death usually ensues within 3–5 years of these.

Clinical features

Aortic stenosis is commonly picked up in asymptomatic patients at routine clinical examination but the three cardinal symptoms are angina, breathlessness and syncope ([Box 18.106](#)). Angina arises because of the increased demands of the hypertrophied LV working against the high-pressure outflow tract obstruction, leading to a mismatch between oxygen demand and supply, but may also be due to coexisting coronary artery disease, especially in old age, when it affects over 50% of patients. Exertional breathlessness suggests cardiac decompensation as a consequence of the excessive pressure overload placed on the LV. Syncope usually occurs on exertion when cardiac output fails to rise to meet demand, leading to a fall in BP.



18.106 Clinical features of aortic stenosis

Symptoms

- | | |
|--|--|
| <ul style="list-style-type: none"> ● Mild or moderate stenosis: usually asymptomatic ● Exertional dyspnoea | <ul style="list-style-type: none"> ● Exertional syncope ● Sudden death ● Episodes of acute pulmonary oedema |
|--|--|

<ul style="list-style-type: none"> ● Angina 	
Signs	
<ul style="list-style-type: none"> ● Ejection systolic murmur ● Slow-rising carotid pulse ● Thrusting apex beat (LV pressure overload) 	<ul style="list-style-type: none"> ● Narrow pulse pressure ● Signs of pulmonary venous congestion (e.g. crepitations)

The characteristic clinical signs of severe aortic stenosis are shown in [Box 18.106](#) . A harsh ejection systolic murmur radiates to the neck, with a soft second heart sound, particularly in those with calcific valves. The murmur is often likened to a saw cutting wood and may (especially in older patients) have a musical quality like the ‘mew’ of a seagull ([Fig. 18.89](#)). The severity of aortic stenosis may be difficult to gauge clinically, as older patients with a non-compliant ‘stiff’ arterial system may have an apparently normal carotid upstroke in the presence of severe aortic stenosis. Milder degrees of stenosis may be difficult to distinguish from aortic sclerosis, in which the valve is thickened or calcified but not obstructed. A careful examination should be made for other valve lesions, particularly in rheumatic heart disease, when there is frequently concomitant mitral valve disease.

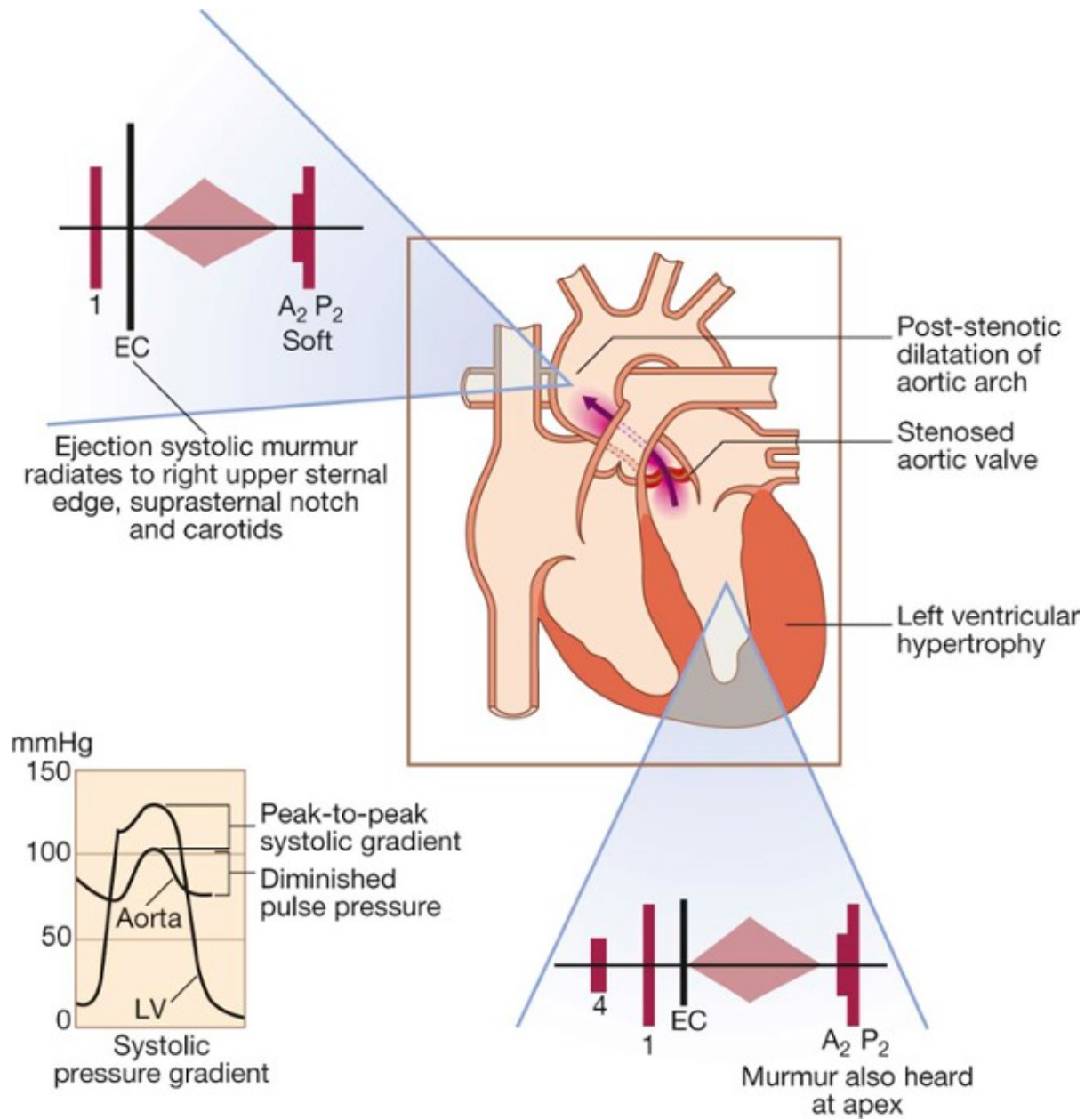


FIG.18.89 Aortic stenosis. Pressure traces show the systolic gradient between LV and aorta. The 'diam...

Investigations

In advanced cases, ECG features of hypertrophy ([Box 18.107](#)) are often gross ([Fig. 18.90](#)), and down-sloping ST segments and T inversion ('strain pattern') are seen in leads reflecting the LV. Nevertheless, especially in old age, the ECG can be normal, despite severe stenosis. Echocardiography demonstrates restricted valve opening ([Fig. 18.91](#)) and Doppler assessment permits calculation of the systolic gradient across the aortic valve, from which the severity of stenosis can be assessed (see [Fig. 18.11](#), p. 536). In patients with an impaired LV, velocities across the aortic valve may be diminished because of a reduced stroke volume, while when aortic regurgitation is present, velocities are increased because of an increased stroke volume. In these circumstances, aortic valve area calculated from Doppler measurements is a more accurate assessment of severity. CT and MRI are useful in assessing the degree of valve calcification and stenosis, respectively, but are rarely necessary.

18.107 Investigations in aortic stenosis

ECG

- Left ventricular hypertrophy (usually)
- Left bundle branch block

Chest X-ray

- May be normal; sometimes enlarged LV and dilated ascending aorta on PA view, calcified valve on lateral view

Echo

- Calcified valve with restricted opening, hypertrophied LV (see [Fig. 18.91](#))

Doppler

- Measurement of severity of stenosis
- Detection of associated aortic regurgitation

Cardiac catheterisation

- Mainly to identify associated coronary artery disease
- May be used to measure gradient between LV and aorta

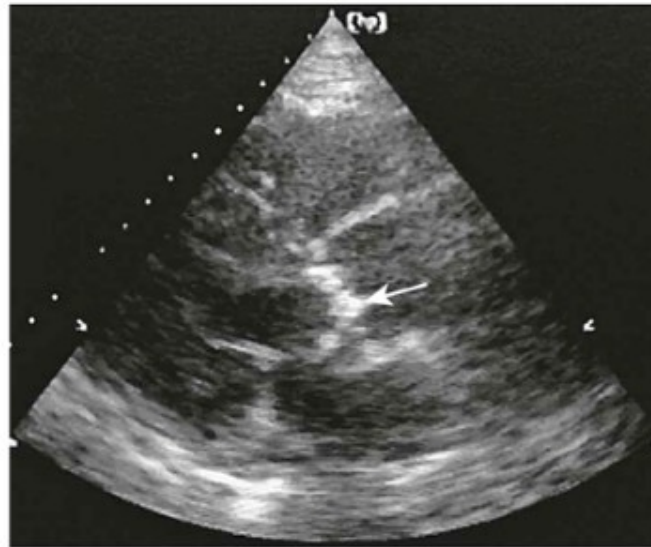


FIG.18.90 **Left ventricular hypertrophy.** QRS complexes in limb leads have increased amplitude with ...





B



C



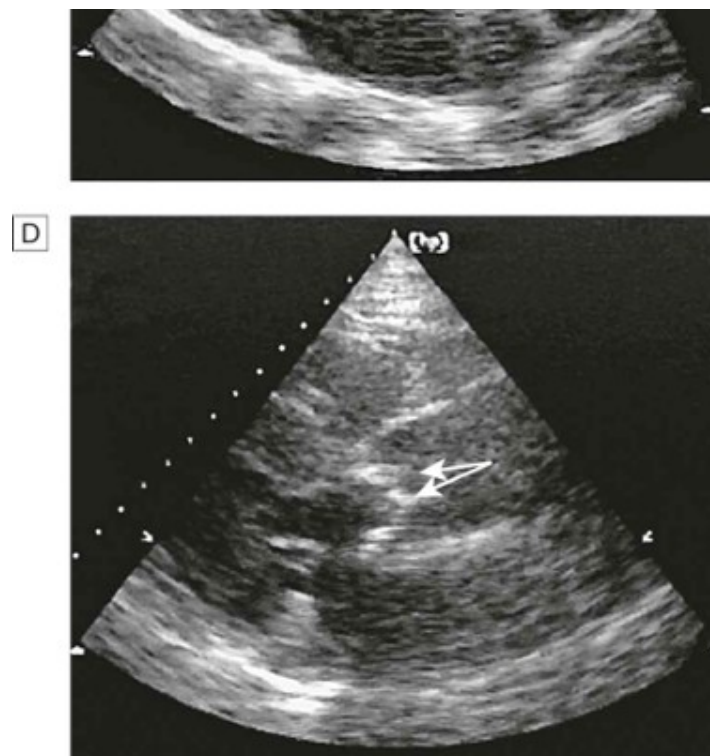


FIG.18.91 Two-dimensional echocardiogram comparing a normal subject with a patient with ca...

Management

Irrespective of the severity of valve stenosis, patients with asymptomatic aortic stenosis have a good immediate prognosis and conservative management is appropriate. Such patients should be kept under review, as the development of angina, syncope, symptoms of low cardiac output or heart failure has a poor prognosis and is an indication for prompt surgery. In practice, patients with moderate or severe stenosis are evaluated every 1–2 years with Doppler echocardiography to detect progression in severity; this is more rapid in older patients with heavily calcified valves.

Patients with symptomatic severe aortic stenosis should have prompt aortic valve replacement. Old age is not a contraindication to valve replacement and results are very good in experienced centres, even for those in their eighties ([Box 18.108](#)). Delay exposes the patient to the risk of sudden death or irreversible deterioration in ventricular function. Some patients with severe aortic stenosis deny symptoms, and if this could be due to a sedentary lifestyle, a careful exercise test may reveal symptoms on

modest exertion. Aortic balloon valvuloplasty is useful in congenital aortic stenosis but is of no value in older patients with calcific aortic stenosis.



18.108 Aortic stenosis in old age

- **Incidence:** the most common form of valve disease affecting the very old.
- **Symptoms:** a common cause of syncope, angina and heart failure in the very old.
- **Signs:** because of increasing stiffening in the central arteries, low pulse pressure and a slow rising pulse may not be present.
- **Surgery:** can be successful in those aged 80 yrs or more in the absence of comorbidity, but with a higher operative mortality. The prognosis without surgery is poor once symptoms have developed.
- **Valve replacement type:** a biological valve is often preferable to a mechanical one because this obviates the need for anticoagulation, and the durability of biological valves usually exceeds the patient's anticipated life expectancy.

Anticoagulants are only required in patients who have atrial fibrillation or those who have had a valve replacement with a mechanical prosthesis.

Aortic regurgitation

Aetiology and pathophysiology

This condition is due to disease of the aortic valve cusps or dilatation of the aortic root (Box 18.109). The LV dilates and hypertrophies to compensate for the regurgitation. The stroke volume of the LV may eventually be doubled or trebled, and the major arteries are then conspicuously pulsatile. As the disease progresses, left ventricular diastolic pressure rises and breathlessness develops.

18.109 Causes of aortic regurgitation

Congenital

- Bicuspid valve or disproportionate cusps

Acquired

- Rheumatic disease
- Infective endocarditis
- Trauma
- Aortic dilatation (Marfan's syndrome, aneurysm, dissection, syphilis, ankylosing spondylitis)

Clinical features

Until the onset of breathlessness, the only symptom may be an awareness of the heart beat ([Box 18.110](#)), particularly when lying on the left side, which results from the increased stroke volume. Paroxysmal nocturnal dyspnoea is sometimes the first symptom, and peripheral oedema or angina may occur. The characteristic murmur is best heard to the left of the sternum during held expiration ([Fig. 18.92](#)); a thrill is rare. A systolic murmur due to the increased stroke volume is common and does not necessarily indicate stenosis. The regurgitant jet causes fluttering of the mitral valve and, if severe, causes partial closure of the anterior mitral leaflet, leading to functional mitral stenosis and a soft mid-diastolic (Austin Flint) murmur.

18.110 Clinical features of aortic regurgitation

Symptoms

Mild to moderate aortic regurgitation

- Often asymptomatic

- Awareness of heart beat, 'palpitations'

Severe aortic regurgitation

- Breathlessness
- Angina

Signs

Pulses

- Large-volume or 'collapsing' pulse
- Low diastolic and increased pulse pressure
- Bounding peripheral pulses
- Capillary pulsation in nail beds: Quincke's sign
- Femoral bruit ('pistol shot'): Duroziez's sign
- Head nodding with pulse: de Musset's sign

Murmurs

- Early diastolic murmur
- Systolic murmur (increased stroke volume)
- Austin Flint murmur (soft mid-diastolic)

Other signs

- Displaced, heaving apex beat (volume overload)
- Pre-systolic impulse
- Fourth heart sound
- Crepitations (pulmonary venous congestion)

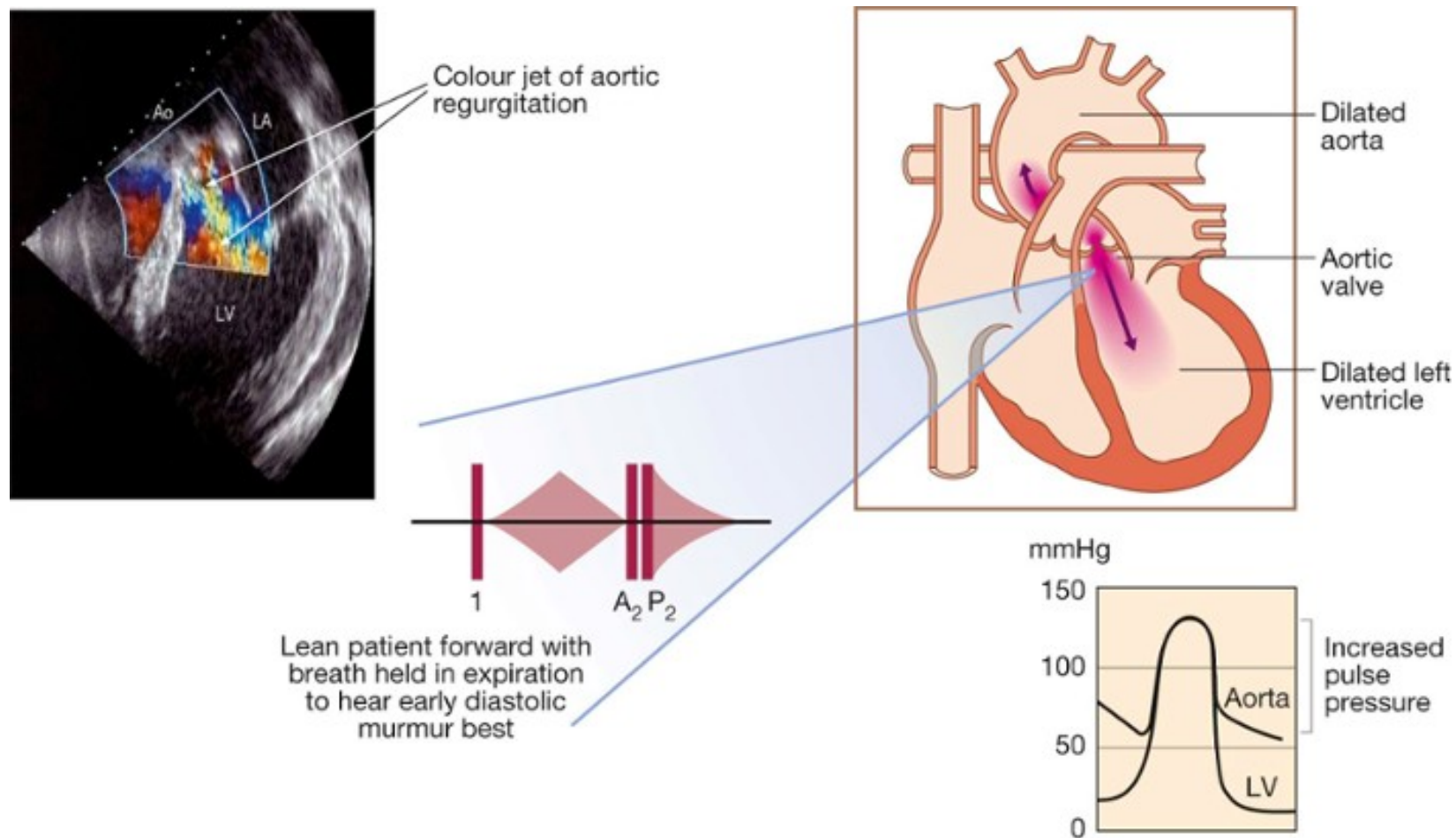


FIG. 18.92 Aortic regurgitation. The early diastolic murmur is best heard at the left sternal edge and m...

In acute severe regurgitation (e.g. perforation of aortic cusp in endocarditis), there may be no time for compensatory left ventricular hypertrophy and dilatation to develop and the features of heart failure may predominate. In this situation, the classical signs of aortic regurgitation may be masked by tachycardia and an abrupt rise in left ventricular end-diastolic pressure; thus, the pulse pressure may be near normal and the diastolic murmur may be short or even absent.

Investigations

Regurgitation is detected by Doppler echocardiography ([Box 18.111](#)). In severe acute aortic regurgitation, the rapid rise in left

ventricular diastolic pressure may cause premature mitral valve closure. Cardiac catheterisation and aortography can help in assessing the severity of regurgitation, and dilatation of the aorta and the presence of coexisting coronary artery disease. MRI is useful in assessing the degree and extent of aortic dilatation.

18.111 Investigations in aortic regurgitation

ECG	
<ul style="list-style-type: none"> Initially normal, later left ventricular hypertrophy and T-wave inversion 	
Chest X-ray	
<ul style="list-style-type: none"> Cardiac dilatation, maybe aortic dilatation Features of left heart failure 	
Echo	
<ul style="list-style-type: none"> Dilated LV Hyperdynamic LV Doppler detects reflux 	<ul style="list-style-type: none"> Fluttering anterior mitral leaflet
Cardiac catheterisation (may not be required)	
<ul style="list-style-type: none"> Dilated LV Aortic regurgitation 	<ul style="list-style-type: none"> Dilated aortic root

Management

Treatment may be required for underlying conditions, such as endocarditis or syphilis. Aortic valve replacement is indicated if aortic regurgitation causes symptoms, and this may need to be combined with aortic root replacement and coronary bypass surgery. Those with chronic aortic regurgitation can remain asymptomatic for many years because compensatory ventricular dilatation and hypertrophy occur, but should be advised to report the development of any symptoms of breathlessness or

angina. Asymptomatic patients should also be followed up annually with echocardiography for evidence of increasing ventricular size. If this occurs or if the end-systolic dimension increases to 55 mm or more, then aortic valve replacement should be undertaken. Systolic BP should be controlled with vasodilating drugs, such as nifedipine or ACE inhibitors. There is conflicting evidence regarding the need for aortic valve replacement in asymptomatic patients with severe aortic regurgitation. When aortic root dilatation is the cause of aortic regurgitation (e.g. Marfan's syndrome), aortic root replacement is usually necessary.